Atrial Arrhythmias
A “Call to Arms” for Congenital Heart Disease Caregivers

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Grow old along with me!
The best is yet to be,
The last of life, for which the first was made:
Our times are in His hand
Who saith, “A Whole I planned,
Youth shows but half; . . .
—Robert Browning, “Rabbi Ben Ezra”1

In his poem “Rabbi Ben (sic Ibn) Ezra,” Robert Browning acknowledges the multitude of physical and nonphysical determinants of life’s richness. His narrator sees these influences as intertwined and spanning the ages from youth through adulthood in timeless fashion. In this week’s issue of Circulation, Bouchardy and colleagues2 determine the prevalence of atrial (tachy)arrhythmias (AA) in adults with congenital heart disease (ACHD) and then analyze the prognostic implications of such. The authors’ findings carry far-reaching implications that, in a fashion that Browning’s narrator would have advocated, extend to principles, design, and application of medical care that begin at the earliest pediatric years and continue throughout the entire development and aging of the ACHD population.

Article see p 1679

Nearly all physicians come into contact with ACHD because current estimates support a prevalence of >1 million adults and nearly 1 million children with congenital heart disease in the United States.3 Analyses of large administrative data sets have emphasized the growing burden (in excess to matched controls) of both cardiovascular and noncardiovascular (diabetes mellitus, renal dysfunction, obesity) disease currently existing in this ACHD population.4 Similarly, all physicians care for patients with AAs, the most common heart rhythm disorder that affects adults. Atrial fibrillation dominates (affecting >23 million Americans, with a prevalence of 0.4% to 5.5%, depending on age) and is a major source of morbidity and mortality because its onset heralds a 4- to 5-fold increase in stroke, a doubling of risk for dementia, a tripling of risk for heart failure, and a 40% to 90% overall increase in mortality.5,6 Over this past decade, AA care has focused on catheter, implantable device, and surgical therapeutic strategies. However, studies such as that by Bouchardy and colleagues that demonstrate the demographics and implications of AA in the ACHD population carry the potential for development of strategies from youth through adulthood that not only prevent the occurrence of AA but may positively affect the overall health of the ACHD population, as well.

To date, few studies have catalogued, analyzed, and then assessed the implications of specific sequelae within the ACHD population at large. Marelli and colleagues7,8 have previously reported their successes in constructing, refining, and analyzing aspects of a province-wide administrative data set that captures all medical interactions for regional inhabitants. Central to their efforts has been accurate and valid definition and recording of entry and outcome variables, as well as analysis of a sufficiently large population of study to allow for an adequate matched control. The current retrospective case-controlled efforts by the authors, although ripe with recognized limitations, serve as a major advance for such study within the field of congenital heart disease care.

In their present study, Bouchardy and colleagues assess the occurrence of atrial tachyarrhythmias (which, by the limitations of their data set, include all atrial flutter, intra-atrial reentrant tachycardia, and both paroxysmal and chronic atrial fibrillation) within the ACHD population who presented for any medical care over an 18-year study period. Coding was accepted only when diagnoses were made by specialty physicians. Prevalence and incidence risks of AA were calculated and correlated with end points of cardiovascular interventions, morbidity, mortality, and a combination of the 3 occurrences. The overall 20-year risk of development of AA was found to be 15% in the ACHD population (up to 3-fold higher than that of age-matched controls), with risk highly correlated to severity of underlying CHD diagnosis. Most striking was the nearly 50% increase in mortality (odds ratio, 1.47; 95% confidence interval, 1.37 to 1.58) noted even within the first year after occurrence of AA, as well as the strength of the relationship between AA and the occurrence of stroke, heart failure, and cardiac interventions. This study does not address whether these relationships were merely associative or causative, but their co-occurrence alone is a call to arms for the ACHD population and its caregivers.

A significant limitation to the control of this study is one that also appears to heighten its import. In calculation of risk, populations were controlled for recognized AA predispositions, defined within the study as presence of hypertension,
coronary artery disease, diabetes mellitus, stroke, heart failure, or recent cardiac surgery. Not controlled by these factors were influences of race, alcohol, or stimulant use, thyroid disease, obesity, presence of metabolic syndrome, valvular heart disease or other existing hemodynamic or autonomic perturbations, known thromboses, hypoxia or other pulmonary disease, sleep apnea, or other inflammatory disease, all of which are recognized as triggers to the development of atrial tachyarrhythmias. Many of these factors have been investigated inadequately within the ACHD population and, in fact, may be overrepresented compared with control populations. These risks may occur over the lifetime of an individual and, of particular import, appear to have the ability to be influenced in preventative fashion.

The major pathological substrates for AA remain a combination of structural, autonomic, and electric (ion channel and gap junction) remodeling that occurs with age and is amplified by other risk factors. Regarding such remodeling in CHD, suture lines and fibrosis conspire together to form the amplified by other risk factors. Regarding such remodeling in CHD, suture lines and fibrosis conspire together to form the foundation for atrial tachyarrhythmia and atrial fibrillation. Although suture lines appear to be a necessary evil occurring in the pursuit of surgical access throughout the heart and vasculature, greater understanding of location-, direction-, and technique-dependent effects of incisions and suturing may affect the development of scar and effects on arrhythmia formation and propagation. Likewise, a focus on the reduction of cardiovascular and noncardiovascular influences on development of elevated atrial pressures, including targeted therapy of valvular abnormalities and aggressive treatment of both diastolic and systolic ventricular dysfunction, may lessen the exacerbation of effects from existing scars on arrhythmia occurrence.

The authors corroborate that increasing CHD lesion complexity (excluding atrial septal defect in the present study) correlates with increasing prevalence of AA. In a separate recent series of 1304 ACHD patients, the prevalence of AA was 10%, with the most important risk factors for AA occurrence being heart failure, advancing age, and lesion complexity. When lesion-specific cohorts were examined, advancing age and physiological abnormalities including heart failure, pulmonary insufficiency, and right atrial size dominated as risk factors. Hope remains that early influences on relief of cyanosis and improving hemodynamics in a lesion-specific fashion may decrease the extent and impact of atrial fibrrosis on development of AA. Such has been witnessed in the early relief of shunting in persons with atrial septal defect, combined accessory pathway ablation and tricuspid valve and right ventricular surgical therapy for the Ebstein anomaly, alternate suture pathways coupled with both ablative techniques and atrial pacing for persons with sinus node dysfunction after atrial switch repair for transposition of the great arteries, and attempts to reduce both the duration and extent of cyanosis as well as to bring suture lines and pressure loading outside of the atrium in the repair of univentricular hearts. Future improvements in quantifying the severity of pulmonary and tricuspid regurgitation may likewise lead to strategies invoking earlier intervention for right-sided obstructive lesions and may carry lesser risk for AA development and improved overall outcomes for affected patients.

Likewise, modern therapy of noncongenital, acquired elevation of atrial pressure currently focuses on improvements in ventricular-vascular coupling, with interventions that in part include reduction of systemic hypertension, obesity control, exercise and fitness training, treatment of metabolic syndrome, reduction of salt and volume excess, and therapy for sleep apnea. Many of these interventions have been pursued less aggressively in the past for the large numbers of ACHD patients who appear affected by such. The present study by Bouchardy and colleagues gives greater reason to further pursue and analyze the outcomes of such interventions.

“Grow old along with me!” Taken alone, Browning’s opening line has been a source of challenge for nearly 150 years, as each reader assesses his or her future and aging. For ACHD patients, the findings in this week’s issue of Circulation confirm that “youth shows but half,” as hemodynamic sequelae, medical disease, and lifestyle changes that are established in youth contribute to the devastation of AA seen in adulthood. CHD medical and surgical clinicians and researchers at all levels, from pediatric to geriatric years, must now be called to the challenge to implement and assess medical interventions and lifestyle changes in these patients. Only by doing this can we restore the statement that “the best is yet to be.”

Disclosures
Dr Landzberg is supported in part by the Dunlevie Foundation.

References
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Circulation. 2009;120:1649-1650; originally published online October 12, 2009; doi: 10.1161/CIRCULATIONAHA.109.899310
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

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